VENOUS VASCULAR MALFORMATION OF LIP – A CASE REPORT

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Abstract

Vascular malformations comprises heterogeneous group of lesions that often present with diagnostic and therapeutic challenge. Here in, we report a case of venous-vascular malformation in the lower lip of an adult male that was locally infiltrative. The lesion was successfully managed with surgical excision and into the one year follow-up, there is no recurrence. It is important for the clinician to recognize the lesion for the appropriate management of this entity.

Key words: Hemangioma, Vascular Malformation, Vascular Tumor

Introduction

Venous vascular malformations are generally defined as a simple, slow flow lesion, and possess an abnormal venous network. They are congenital lesion but clinically become more obvious during the adolescent stage.1 Most often they are improperly diagnosed due to confusion in terminology with misnomers such as hemangioma, cavernous hemangioma, phlebangioma and phlebangiomatosis that often results in improper management. In this article, we describe a case of venous malformation in the adult male in the lower lip which was successfully treated surgically.

Case Report

A 21 year old male patient presented with the chief complaint of painless, gradually progressive swelling in the lower lip of six month duration. Clinical examination revealed a sub mucosal swelling of size approximately 1x1 cm in diameter. It was bluish in color with smooth surface and rubbery in consistency. [Figure-1A] The lesion was not freely mobile and mildly tender on palpation. No evidence of palpable thrill was noticed.

Based on the distinctive clinical appearance of the lesion a provisional diagnosis of low flow vascular (venous) malformations was made with the differential diagnosis of blue nevus and other soft tissue tumors.

Surgical excision of the lesion was planned under local anaesthesia. Superficial mucosal incision was made and subsequent reflection of the mucosa revealed a soft, dark brownish multiple nodules, infiltrating the adjacent muscle fibers [Figure-1B]. The lesion was dissected carefully from the muscle fibers and excised as a whole from the surrounding tissue [Figure-1C]. Grossly the specimen appeared to be solitary mass with multiple nodules, dark brownish in color, smooth surface and measured about 1x1 cm and soft in consistency. [Figure-1D]

Histopathological examination of the hematoxylin and eosin (H&E) stained sections basically showed multiple, thin walled, large vascular channels filled with RBCs in the fibro-cellular connective tissue stroma. [Figure-2A&B] The thin vascular lining showed spindle endothelial cells. [Figure-2C&D] Normal striated muscle fibers in the fibrous stroma were also seen. Based on the clinical and histopathological features, a final diagnosis of venous vascular malformation was made.

Discussion

Vascular malformations by definition are congenital anomalies and are thus present at birth although clinically may not be evident at the time of birth.1

Figure 1: Sub mucosal bluish nodule in the left side lower lip (A), lesion after surgical exploration (B), surgical site after complete excision of the lesion (C) and gross specimen (D)

Figure 2: Large vascular (venous) channels filled with RBCs in the connective tissue stroma (A&B) (H&E4X) and thin vascular lining comprising endothelial cells (C&D) (H&E 40X)
Mulliken and Glowacki (1982) gave most useful and widely accepted classification of vascular anomalies. They had classified vascular anomalies as either vascular tumor with proliferating endothelial cells (hemangioma) or vascular malformations due to an abnormal embryonic development (capillary, venous, lymphatic, arterial, arterio-venous and malformations). Their classification scheme was based on mainly the clinical, histologic, histochemical and biochemical features. Also vascular malformations are classified as slow flow malformations (capillary, venous, lymphatic, capillary-venous, capillary-venous-lymphatic malformations) and high flow malformations (arteriovenous fistula, arteriovenous malformations) which is highly useful in the clinical management of the lesions.

The pathogenesis of vascular malformations involves loss of ability to secrete platelet derived growth factor (PDGF) and transforming growth factor β (TGF β) by endothelial precursor cells at the early stage of embryogenesis, resulting in failure of recruitment of adventitial cells surrounding developing vessels leading to formation of structurally unsupported vessels. In most of the cases, the clinical manifestation is generally delayed till the prepubertal stage, due to insufficient pressure in the cardiovascular circulation. After prepubertal stage, maturation of cardiovascular system results in increased systemic pressure, leading to expansion of structurally weak vessels in the affected individuals.

Clinically venous malformations are characterized by a soft, non-compressible, non-pulsatile tissue mass. The overlying skin usually appears blue, although normal sometimes. The most common locations are head and neck (40%), extremities (40%) and trunk (20%) of cases. Unlike hemangioma, vascular malformations grow continuously and do not regress. Exacerbations of growth may be seen during puberty and pregnancy (hormonal influence) or due to trauma, infection, thrombosis.

Although most cases occur subcutaneously, lesions involving deeper tissue plane (muscle, abdominal viscera, and bone) is not rare. The severity of symptom mainly depends on individual lesion size and location. Deep cutaneous and intramuscular lesions cause discomfort with exertion. Large oral lesion can potentially cause bleeding, distortion of dentition, speech difficulties and airway obstruction.

Different imaging modalities play a very crucial role in the diagnosis and management of vascular malformations. Conventional radiography demonstrates a soft tissue mass and occasionally the phleboliths within the lesion. Doppler ultrasound examination is most helpful in differentiating the low flow lesions such venous malformation from lesions with high flow characteristics e.g. arterial malformation, arterio-venous (A-V) malformation. Venous malformations generally present as hypoechoic or heterogeneous lesions with mono-phasic, low velocity flow. Absence of flow may be seen in thrombosis or in very small lesions (equipment inability to detect the flow).

InComputed tomography (CT) scan vascular malformations present as hypoattenuating or heterogeneous lesions and shows slow peripheral expansion with the injection of contrast material. Although CT scan demonstrate the lesion extension, superior delineation is possible with magnetic resonance (MRI) imaging. With CT scan more clear depiction of phlebolith is possible.

Excellent delineation of lesion extent and its relation with adjacent structures is possible with MRI scan and it remains the investigation of choice in the diagnosis and management of vascular malformations. Venous malformations, in T1 weighted MR imaging generally show hypo-intense or iso-intense signal. Heterogeneous signal intensity is possible in situation such as hemorrhages or thrombosis within the lesion. At T2 weighted imaging venous malformation usually show bright signal intensity. T2 weighted images more clearly depicts the extension of lesion into the adjacent structures. Although MRI is most sensitive in the diagnosis and superior in delineating the extent of venous malformations, its findings must always be correlated with the clinical and Doppler US features to make the final diagnosis. In atypical cases, direct percutaneous phlebography is essential for confirmation of diagnosis.

Sclerotherapy, surgery and laser therapy are the most common options used in the management of venous malformations. Absolute ethanol is the most commonly used sclerosing agent. Also alcoholic solution of zein (a corn protein) or a mixture of zein, alcohol and contrast medium is also found to be more effective. Fistulization is the common complication of using zein based solutions due to extrusion of injected materials. Sodium tetradecyl sulphate is another agent which is mainly used in oromucosal or more superficial cutaneous lesions, since the risk of tissue necrosis is negligible. Superficial tissue necrosis, neural toxicity (mainly with alcohol) is the most common complication associated with sclerotherapy. Systemic adverse effects are very rare and include hemolysis with renal toxicity and cardiac arrest. The injection of sclerosing agents induces the inflammatory reaction, which may result in aggravation of pain in the individuals. Non-steroidal anti-inflammatory drugs or corticosteroids are beneficial in reducing the severity of symptoms.

Surgical excision of the lesion is generally done, when sclerotherapy is incomplete or for small lesions in accessible locations such as cutaneous and oro-mucosal region.

Photocoagulation using argon or neodymium, yttrium, aluminium garnet laser is mainly used in superficial lesions and for deep lesions can be managed with subcutaneous insertion of laser probes. Satisfactory results with minimal scarring have been reported but recurrences are common.

After proper clinical diagnosis, the current case was successfully managed with surgical excision alone, since it was smaller in size and in superficial location, avoiding complication associated with sclerotherapy. The post
operative period was uneventful and one year into the follow no recurrence was noticed.

References


